

Calciophylaxis, a rare disease with fatal outcome

Noman Ahmed Jang Khan, Hafiz Umair Siddiqui, Muhammad Asif, Muhammad Arslan Karim

Madam, Calciophylaxis which is also known as calcific uraemic arteriopathy (CUA) is a poorly understood syndrome of vascular calcification and skin necrosis. Skin lesions develop suddenly and progress rapidly. Most commonly the lesions appear on the lower extremities. The lesions are typically erythematous plaques or papules with central cutaneous necrosis.¹ The pathogenesis of calciophylaxis is obscure. Disorders that are implicated in the pathogenesis of calciophylaxis include chronic renal failure, obesity, hypercalcaemia, hyperphosphataemia, hypothyroidism, hyperparathyroidism and perhaps a variety of hypercoagulable states. Imbalance of many molecular factors including bone morphogenic protein-2 (BMP-2), osteopontin, osteoprotegerin, parathyroid related protein (PTHrP), matrix-glycoprotein and phosphate have been studied.²

Serum calcium, phosphate, blood urea nitrogen (BUN), creatinine, parathyroid hormone levels, coagulation factors profile, prothrombin time, activated partial thromboplastin time, protein C and S assay, erythrocyte sedimentation rate (ESR), c-reactive protein (CRP), amylase and lipase can help establish a presumed cause. To exclude vasculitis perinuclear anti-neutrophil cytoplasmic antibody (p-ANCA), cytoplasmic antineutrophil cytoplasmic antibodies (c-ANCA) and anti neutrophil antibodies titres can be obtained. Plain x rays can show calcification of soft tissues. A deep skin biopsy with subcutaneous adipose tissue is necessary to confirm diagnosis and the most common histopathological finding is a septal calcifying panniculitis.²

Early diagnosis of calciophylaxis and multi-disciplinary approach can result in improvement. Aggravating

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MBBS, Dow Medical College, Karachi, Pakistan.

Correspondence: Noman Ahmed Jang Khan.

Email: noman_jang020@hotmail.com

conditions should be addressed and trigger factors be eliminated. In patients with end stage renal disease, increasing the frequency of dialysis can ameliorate the symptoms. An aggressive programme of wound care and adequate pain control should be initiated. Some studies have shown proven benefit of hyperbaric oxygen in the healing of wounds.³ In patients with elevated calcium and phosphate levels, calcium and phosphate products may be lowered to less than 55 mg/dl². Cinacalcet, a calcinomimetic can be used as an alternative to surgery for hyperparathyroidism.⁴ For patients with medically refractory hyperparathyroidism, parathyroidectomy is recommended.⁴ Bisphosphonates have some role in the management of calciophylaxis associated with increased calcium and phosphate. Intravenous sodium thiosulphate, an antioxidant is increasingly being used as treatment of choice for calcific uraemic arteriopathy.⁴

The mortality rate of calciophylaxis is as high as 60-80%.⁵ The most common cause of death is sepsis from infected, necrotic skin lesions. The high mortality rate of calcific uraemic arteriopathy warrants appropriate treatment strategy and risk factors control. The preventive aspect of this disorder should be given pivotal importance as this is the most effective way to combat calciophylaxis.

References

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